Primary Idiopathic Chylopericardium:  
Report of a case and review of the literature

Young Sup Yoon, Won Heum Shim, Tae Sub Chung and Young Sik Lee

Primary or isolated chylopericardium of unknown etiology is considered a rare cause of pericardial effusion. Its etiology is obscure but certain communication between the lymphatic system and pericardial sac was suggested. Up to 1991, there was only one case report that successfully showed the direct communication by a lymphangiogram. We report a case of chylopericardium occurring in a nearly asymptomatic 22-year-old man with no apparent history of trauma, infection or mediastinal neoplasms, in which we succeeded in visualizing the communication between the thoracic duct and pericardial sac by lymphangiography and computed tomography of the chest. A review of the previous cases is described also.

Key Words: Primary idiopathic chylopericardium, lymphangiography

Primary chylopericardium of idiopathic etiology is a very rare clinical entity. About thirty cases were previously reported in the medical literature after Madison and Logue (1957) reported the first one. The case of primary chylopericardium, however, was first described by Groves and Effler (1954), which was associated with cystic hygroma in the mediastinum. As the name suggests, chylous effusion in the pericardium should be causally related with the lymphatic system. Many previous investigators tried to show the direct communication between the pericardial sac and thoracic duct or other lymphatic tributaries within the chest, but only one was able to demonstrate it (Mask et al. 1990). However, many previous cases of the chylopericardium of diverse etiology were cured by thoracic duct ligation, which suggests leaks from thoracic lymphatics should have resulted in the chylos effusion. We experienced a case of primary idiopathic chylopericardium in a 22-year-old man, who had asymptomatic massive pericardial effusion. It was falsely treated as tuberculous pericarditis for three years. After it was confirmed as chylos effusion by pericardiocentesis, we took radiologic studies and could demonstrate clear communication between the pericardial space and thoracic duct using a lymphangiography and computed tomography during lymphangiography of the chest. A review of the previous cases with diagnostic and therapeutic options will be described.

CASE

A 22-year-old man was admitted to the division of cardiology at Yonsei Cardiovascular Center, Yonsei University with the diagnosis of "cardiomegaly". He was in his usual state of good health until April, 1989 when he first felt mild substernal discomfort. His past history revealed that the cardiomegaly had been discovered incidentally on his routine chest X-ray film upon entering military service in 1988. But
at that time he didn't visit any hospital for further evaluation because he had been asymptomatic. There was no history of tuberculosis or chest trauma.

On admission he complained of mild dyspnea on exertion and a mild cough. The temperature was 36.8°C, the pulse rate 90 per minute and the blood pressure 130/90 mmHg without pulsus paradoxus. On physical examination he appeared fatigued but seemed comfortable at rest. Neck veins were not distended. The breathing sound was clear. The heart sounds were easily heard and murmurs were not audible; pericardial friction rub was not audible. No organomegaly or tenderness was found in the abdomen. All peripheral pulses were easily palpable. There was no peripheral cyanosis, clubbing or edema. The roentgenogram of the chest showed marked enlargement of the cardiac shadow with a globar shape. Two dimensional echocardiographic examination revealed a massive pericardial effusion and diastolic collapse of the right ventricle. An electrocardiogram demonstrated a sinus rhythm, with low voltage QRS complexes in the limb leads. The blood count, serum electrolytes, blood chemistry values and urinalysis were normal. His evaluation was negative for viral, bacterial and autoimmune causes of the fluid collection. PPD skin test was positive in 5TU but repeated sputum study for tuberculosis was negative. There seemed to be no apparent cause for the enlargement of the cardiac shadow. On the 3rd hospital day, a left anterior thoracotomy was performed because the pericardial effusion gradually progressed and no possible cause was detected. The pericardial sac was enlarged and distended without evidence of inflammation. Incision of the pericardium resulted in the immediate gushing out of 800 mL of milky-white pericardial fluid. After evacuation of the pericardial fluid, the heart seemed to be normal in size, and the epicardium was coated with a thin milky fluid. There was no evidence of neoplasm, tuberculosis, or other pathologic conditions. The pericardium revealed grossly normal mucosa, with no evidence of inflammation on the heart surface. A small biopsy disk of the pericardium was excised. At the time of operation, the surgeon thought that an unusual form of tuberculous pericarditis was the most probable diagnosis. A soft rubber tube was inserted into the pericardial sac and was exterio-

rized through a separate wound. After pericardial drainage, his symptoms improved markedly and follow-up echocardiographic examination revealed that minimal pericardial effusion was left. Routine examination of the pericardial fluid revealed no tumor cells: the specific gravity was more than 1.035; the pH was 7.6; the protein was 7.6 g/dl; the glucose was 103 mg/dl; LDH was 105 IU/dl. But cholesterol and triglyceride levels were not quantified. There were 10,000 RBCs/mm³ and 4,600 WBCs/mm³ with 100% of mononuclear cells. Routine bacterial and tuberculous cultures were subsequently reported as negative. The adenosine deaminase level was 1.8 U/l. Considering these clinical and laboratory findings, nonspecific chronic pericarditis with effusion was our clinical diagnosis. The pericardial tube was left in place for 4 days on dependent drainage during which time 350 mL of pericardial fluid was collected. The tube was removed and the wound healed by primary intention one week later. The pericardial biopsy, later confirmed, disclosed only fibrous thickening without evidence of inflammation. At that time, we decided to try the anti-tuberculous medication-Streptomycin 1.0 g IM, Isoniazid 400 mg PO, Rifampicin 600 mg PO, Ethambutol 800 mg PO-for both diagnostic and therapeutic purpose. He was discharged 3 weeks after the operation. After discharge he was regularly followed up at a private clinic while continuing on the anti-tuberculous medication. In July 1991, he revisited our hospital because of a reaccumulation of the pericardial fluid diagnosed at the private clinic. According to him his cardiac silhouette had been progressively enlarged 6 months after discharge. The anti-tuberculous medication had been continued without stopping. Echocardiography performed at an outpatient clinic revealed a large amount of pericardial effusion without evidence of hemodynamic compromise. The ejection fraction was 58% and no regional wall motion abnormality was seen. Because of a benign clinical course and insufficient treatment time, it was decided that antituberculous medication was to be continued with close observation. 6 months later, a thyroid function test performed at an outpatient clinic revealed a euthyroid state-3.35 μIU/ml of TSH, 170.57 g/ml of T3 and 7.65 μg/ml of T4. Echocardiography performed in April, 1992 disclosed no significant interval change of the
amount of pericardial effusion (Fig. 1). Therefore, he was readmitted to our hospital in May, 1992 for further evaluation. He only complained of general weakness without chest discomfort or dyspnea. On admission he was afebrile. The pulse rate was 84 per minute and the blood pressure was 130/80 mmHg. On physical examination neck veins were not distended and the peripheral pulses were normal. The breathing sound was clear without rale. The heart sounds were slightly decreased and pericardial friction rub was clearly audible. The hemoglobin was 15.0 g/dl, the hematocrit 48%, and the white-cell count 5,300/mm³. The platelet count was 118,000 /mm³ and the ESR was 13 mm/hr. Blood chemistry showed that cholesterol was 143 mg/dl, triglyceride 68 mg/dl, HDL-cholesterol 37 mg/dl, total protein 7.6 g/dl, albumin 3.7 g/dl, urea nitrogen 9.5 mg/dl, creatinine 0.9 mg/dl, SGOT (AST) 12 IU/l, SGPT (ALT) 10 IU/l and LDH 99 IU/l with a normal isoenzyme pattern. The prothrombin time was 13.9 seconds with a control of 10.8 seconds (INR 1.12). The urinalysis was normal. Antinuclear antibody, anti DNA, LE cell and rheumatoid factor were all negative. The complement level and β-microglobulin level were within normal range. Cold ag.

Fig. 2. Pericardial fluid drained from pericardial sac shows milky-yellowish appearance suggesting fat content within the fluid.
glutinin and Anti-HIV were negative. ASO titer was 200 Todd units.

On the 2nd hospital day pericardiocentesis with subxiphoid tube drainage was performed. 500 ml of milky yellowish fluid was drained through the draining tube immediately (Fig. 2). Examination of the pericardial fluid showed a specific gravity of 1.035, pH 7.4, protein 6.7 g/dl, glucose 116 mg/dl, amylase 34 U/dl, LDH 337 IU/l, total cholesterol 106 mg/dl, triglyceride 858 mg/dl, RBC 200/mm³, WBC 250/mm³, with 100% of mononuclear cells. The adenosine deaminase level was 17.3 U/l at 37°C and CEA level was 10.5 ng/ml. Routine cultures for bacteria and tuberculosis were reported as all negative. Cytologic examination showed no tumor cells. 50 to 100 ml of pericardial fluid drained daily through the pericardial tube. The color of the drained fluid showed some interesting variations after meals. The color was usually a clear serous tint, but about one hour after each meal it turned milky-white. The drained pericardial fluid cleared upon the addition of ether and a Sudan-Black B stain showed fat droplets in the fluid by polarized microscopy. These findings with inappropriately elevated triglyceride in the pericardial fluid sufficed to diagnose primary idiopathic chylopericardium. On the 12th hospital day the patient underwent thoracotomy. With the use of the subxiphoid approach a pleuropericardial window was made exposing the pericardium extrapleurally with the insertion of the draining tube. No lymphatic communication with the pericardial space could be found and no definite thoracic duct structure could be identified at the level of the pericardium. The pericardial sac showed no gross thickening and a milky-yellowish effusion was found after opening the pericardial sac. After removing much of the pericardial fluid, a large pericardial window of 6×6 cm size and a pericardiopleural fistula with left hemithorax was made and a 32Fr thoracic catheter was placed in the left pleural cavity in an attempt to prevent pericardial fluid collection and cardiac tamponade. The histologic examination of

**Fig. 3. Direct lymphangiography shows the normal passage of the thoracic duct within the chest draining into the subclavian vein and another aberrant contrast flow at the mid-portion of the thoracic spine.**
the biopsied specimen showed marked fibrous thickening and spotted calcification with evidence of chronic inflammation. Computed tomography of the chest revealed no evidence of mediastinal lymphadenopathy, anomalies or tumor mass.

To find any anomalous communication between lymphatic channels and the pericardial sac, a direct lymphangiography was performed by a slow injection of lipiodol into a cannulated lymphatic vessel of the left foot. Serial films were taken of the lower extremities, pelvis, abdomen and chest. The lymphangiogram showed a normal progression of the contrast medium up to the diaphragm. No abnormalities were found in the lower extremities or intra-abdominal lymphatic vessels. But at the level of the midportion of the thoracic spine, shallow spotted and linear contrast pooling was seen anterior to the esophagus between the thoracic duct and posterior portion of the left atrium (Fig. 3, 4). But much of the contrast media from the thoracic duct drained into the left subclavian vein. To confirm the leakage and the exact anatomic structure of the anomalous communication, we took a computed tomography of that lesion immediately. It clearly showed a linear contrast-filled structure suggesting abnormal mediastinal lymphatics between the thoracic duct and left posterior part of the pericardium which caused chylous effusion in the pericardial sac (Fig. 5). One week after removing the drainage tube, an echocardiogram showed minimal pericardial effusion. So we concluded that the pericardio-pleural fistula was enough to drain the newly formed chyle in the pericardium. On the 7th day after operation he was discharged from the hospital on a low fat diet.

Three months after discharge, follow-up echocardiography showed that pericardial effusion recurred in a moderate amount. But he felt no symptoms and enjoyed normal life. Only pericardiocentesis was performed with tube drainage for 4 days. The pericardial fluid analysis showed the same nature as the previous one. He returned to full-time work and activities on the follow-up time thereafter. Four months later, no further pericardial effusion was detected on the chest X-ray. If reaccumulation is detected later, we will perform the surgical ligation of the thoracic duct.
DISCUSSION

The accumulation of chyle in the pericardial space, therefore called chylopericardium, is an unusual clinical entity. The primary chylopericardium is even rarer. Isolated or primary chylopericardium was first reported in 1888 by Hasebrock who described 22.6 ml of chylous fluid removed at an autopsy from a patient who died of aspiration secondary to tracheal stricture and ulceration (Hasebrock, 1888). After that, three cases of chylopericardium associated with chylothorax were reported in the review of 100 cases of nontraumatic chylothorax (Yater, 1935). The clinical entity of “primary chylopericardium” was first described by Groves and Effler who used the term to describe an isolated recurrent accumulation of chyle in the pericardium associated with cystic hygroma in the mediastinum (Groves and Effler, 1954). According to our review of the medical literature, there have been about sixty prior reports of primary chylopericardium, among which thirty cases are idiopathic in origin (Table 1).

Table 1. Summary of symptoms and treatment in reported cases of primary idiopathic chylopericardium

<table>
<thead>
<tr>
<th>References</th>
<th>Year</th>
<th>Sex/Age (yr)</th>
<th>Symptom</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Madison and Logue</td>
<td>1957</td>
<td>F/40</td>
<td>exertional dyspnea</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Bartel and Neute</td>
<td>1964</td>
<td>M/8</td>
<td>symptom free</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Knight</td>
<td>1965</td>
<td>M/62</td>
<td>dyspnea, cough</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Hudspeth and Miller</td>
<td>1967</td>
<td>M/22</td>
<td>symptom free</td>
<td>TDL and resection</td>
</tr>
<tr>
<td>Yakopoulos et al.</td>
<td>1967</td>
<td>M/40</td>
<td>symptom free</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Fawal et al.</td>
<td>1967</td>
<td>M/36</td>
<td>epigastric discomfort</td>
<td>TDL, hemipherecardiectomy</td>
</tr>
<tr>
<td>Daniel and Bressie</td>
<td>1969</td>
<td>M/38</td>
<td>cough, substernal pain</td>
<td>TDL and resection</td>
</tr>
<tr>
<td>Rouchu et al.</td>
<td>1969</td>
<td>F/32</td>
<td>symptom free</td>
<td>TDL, hemipherecardiectomy</td>
</tr>
<tr>
<td>Ketelers et al.</td>
<td>1972</td>
<td>M/15</td>
<td>symptom free</td>
<td>TDL, hemipherecardiectomy</td>
</tr>
<tr>
<td>Puig-Massana et al.</td>
<td>1972</td>
<td>M/12</td>
<td>symptom free</td>
<td>TDL, hemipherecardiectomy</td>
</tr>
<tr>
<td>Csanady and Kovacs</td>
<td>1973</td>
<td>F/26</td>
<td>dyspnea, edema</td>
<td>hemipherecardiectomy</td>
</tr>
<tr>
<td>Patney</td>
<td>1974</td>
<td>M/19</td>
<td>symptom free</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Dunn</td>
<td>1975</td>
<td>M/11</td>
<td>fatigue</td>
<td>TDL</td>
</tr>
<tr>
<td>Savran et al.</td>
<td>1975</td>
<td>M/29</td>
<td>symptom free</td>
<td>Pericardiotomectomy</td>
</tr>
<tr>
<td>Motoji et al.</td>
<td>1975</td>
<td>F/14</td>
<td>symptom free</td>
<td>Division of thoracic duct</td>
</tr>
<tr>
<td>Charniels et al.</td>
<td>1977</td>
<td>F/18</td>
<td>symptom free</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Sakauki et al.</td>
<td>1977</td>
<td>M/43</td>
<td>fatigue</td>
<td>TDL</td>
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<td>Nagamura et al.</td>
<td>1978</td>
<td>F/23</td>
<td>palpitation, dyspnea</td>
<td>TDL, window</td>
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<td>1978</td>
<td>M/10</td>
<td>cough</td>
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<td>Suzuki et al.</td>
<td>1978</td>
<td>F/30</td>
<td>fatigue</td>
<td>TDL, window</td>
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<td>Ross et al.</td>
<td>1979</td>
<td>M/19</td>
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<td>Chon et al.</td>
<td>1979</td>
<td>M/3mo*</td>
<td>cough</td>
<td>TDL, window</td>
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<td>Kimura et al.</td>
<td>1979</td>
<td>M/14</td>
<td>symptom free</td>
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<td>Harada et al.</td>
<td>1982</td>
<td>F/7</td>
<td>symptom free</td>
<td>pericardiotomectomy</td>
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<tr>
<td>Sheidt et al.</td>
<td>1986</td>
<td>M/19</td>
<td>symptom free</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Mask et al.</td>
<td>1990</td>
<td>F/37</td>
<td>exertional dyspnea</td>
<td>TDL</td>
</tr>
<tr>
<td>Musemeche et al.</td>
<td>1990</td>
<td>M/12</td>
<td>symptom free</td>
<td>TDL, window, pericardiotomectomy</td>
</tr>
<tr>
<td>Bendayan et al.</td>
<td>1991</td>
<td>M/26</td>
<td>symptom free</td>
<td>pericardiotomectomy</td>
</tr>
<tr>
<td>Matsuda et al.</td>
<td>1991</td>
<td>F/8</td>
<td>symptom free</td>
<td>TDL, window</td>
</tr>
<tr>
<td>Itoh et al.</td>
<td>1991</td>
<td>M/79</td>
<td>symptom free</td>
<td>No treatment</td>
</tr>
</tbody>
</table>

TDL: thoracic duct ligation
Window: pericardial window formation
Pericardiotomy: including partial pericardiotomy and hemipherecardiectomy
*: month
Idiopathic chylopericardium which literally means it has no known etiology, usually arises from congenital lymphangioma, mediastinal neoplasms including hamartoma, cystic hygroma, thrombosis of the subclavian vein, nonsurgical trauma, radiation, or thoracic and cardiac surgeries (Bessone et al. 1971; Thomas and McGoon, 1971; Dunn, 1975; Harada et al. 1982; Rose et al. 1982; Bhatti et al. 1985; Tchervenkov and Dobell, 1985). Unlike chylopericardium, primary idiopathic chylopericardium should be diagnosed after exclusion of the known causes for accumulation of chyle in the pericardial sac. The main factors inducing chylopericardium include mechanical obstruction of the thoracic duct or its lymphatic tributaries, or disturbances of the lymphatic drainage into subclavian veins which result from surgical or traumatic injury or lymphatic blockage by neoplasm, tuberculosis, or congenital lymphangiomatosis. As we guess form its terminology, the pathophysiology of idiopathic chylopericardium remains obscure. But “chyloous pericardium” implies the presence of a fistula between the lymphatic system coming from the abdomen and the pericardial sac at the same point. It other words, chylopericardium means a communication between the pericardial sac and the thoracic duct carrying chyle. In fact, some investigators proposed the existence of the direct microscopic connections between the pericardial sac and the thoracic duct to account for chylopericardium (Bartel and Neute, 1964). In idiopathic chylopericardium, factors in the accumulation of chyle in the pericardial space were suggested as follows: (1) obstruction of the thoracic duct with a resultant increase in pressure-but lymphatic blockage would not necessarily result in reflux and chylopericardium since other collaterals are possible (Lee, 1922; Blalock et al. 1937; Csanady and Kovacs, 1973; Dunn, 1975), (2) failure to establish collateral lymph drainage to the right thoracic duct or lymphaticovenous connections to relieve pressure, (3) reflux of chyloous lymph through normal lymphatic channels or through channels lacking valves, which drain the pericardium and the heart (Dunn, 1975). In fact, many previous investigators tried to show the direct communication by radiologic methods or by open thoracotomy. While anomalies and obstruction of the thoracic duct have been demonstrated by these methods, visualization of the direct communication was unsuccessful except for a pooling of the contrast within the pericardial area (Goldstein et al. 1969; Yoshida et al. 1969; Chavez et al. 1973; Bhatti et al. 1985). The demonstration of direct communication between the thoracic duct and pericardial space was achieved by Mask and coworkers (1990). They clearly visualized the leaking flow of contrast media from the thoracic duct to the pericardial sac by lymphangiography and computed tomography of the chest. In our case, we also succeeded in the visualization of the linear contrast leakage from the thoracic duct at the midpoint of the thoracic duct into the upper and posterior part of the pericardium using direct lymphangiography and computed tomography of the chest. However, because the nature of the communication remains obscure in many of the other previous cases, further clinical and experimental investigation would be necessary to clarify the pathophysiology of primary idiopathic chylopericardium.

Idiopathic chylopericardium has been documented from infants to 79 year olds and there is no age preference. The sex distribution was nearly equal between males and females. In most cases, duration of the enlargement of the cardiac silhouette was more than 1 week, which suggested the chronic nature of the pathophysiologic mechanism. Considerable cases found in the adult life suggest that chylopericardium had been present for more than one year, even upto 15 years, at the time the patients were studied (Dunn, 1975; Harada et al. 1982; Mask et al. 1990; Itoh et al. 1991). In our case, it was falsely treated as tuberculous pericarditis for 3 years. But in other chylopericardium, it was also common to be treated as tuberculous or other pericardial effusions before the correct diagnosis was made. Thus primary chylopericardium should be considered among the differential diagnosis of chronic pericardial effusion. Although this disease is very rare, an awareness of this entity should lead to prompt diagnosis because it has simple characteristics. The appearance and symptomatology associated with this disease include fatigue, dyspnea, upper abdominal discomfort, cough, chest pain, and palpitation. But in about half the cases the patient had no symptoms. Asymptomatic or mild symptomatic patients associated with an unexplained enlargement of the cardiac silhouette on a chest roentgeno-
gram was a typical feature of this disease. Rarely, severe respiratory difficulty or acute cardiac tamponade was reported in both postoperative and primary chylopericardium (Miller et al. 1958; Jacob et al. 1974; Bakey and Wijers, 1984; Lee et al. 1987; Pereira, 1988). On physical examination, there may be cardiomegaly by percussion, distant heart sounds on auscultation, evidence of cardiac tamponade, neck vein distension, hepatomegaly, pulsus paradoxus, gallop rhythm, and pericardial friction rub. This disorder can cause some complications such as cardiac tamponade that may result in sudden death, constrictive pericarditis and a loss of lymph fluid which leads to impingement and aberration of imposing structures and their function (Morishita et al. 1985; Mask et al. 1990). One characteristic feature of our case is that the draining fluid through the pericardial or mediastinal tube was usually serous but it became opalescent and milky about an hour after meals, which strongly suggested some relation between feeding and drainage fluid.

The diagnostic modalities essential to evaluate suspected chylopericardium include the following: (1) chest X-ray with special emphasis on the cardiothoracic ratio, (2) M-mode or two-dimensional echocardiography which easily and accurately reveals pericardial effusion, (3) computed tomography of the chest which reveals a density compatible with fat in the pericardial sac and other lymphadenopathy or which excludes other structural diseases in the chest, (4) the transthoracic pericardiocentesis and pericardial fluid analysis which is inevitable to confirm the diagnosis of the chylopericardium. The characteristics of the fluid are a milky-white appearance which clears promptly after addition of ether, presence of fat droplets demonstrated by a Sudan III stain, relatively high triglyceride content compared with cholesterol, high protein content and predominance of the lymphocytes. (5) other diagnostic tests that attempted to demonstrate communication between lymphatic channels and the pericardial sac are as follows. Administration of Sudan III-stained corn oil orally and confirmation of the presence of dye in the pericardial draining fluid was a useful technique (Grove and Effler, 1954; Madison and Logue, 1957; Miller et al. 1959). Another method was oral administration of 131-I-labeled triolen followed by analysis of the pericardial fluid versus blood for radioactivity or external cardiac imaging (Hudspeth and Miller, 1966; Yankopoulos et al. 1967). Recently the most commonly used method has been direct lymphangiography or radionuclide lymphangiography which has a goal to demonstrate communication between lymphatic vessels and thoracic structures directly. But frequently these trials have failed to visualize it. Successful lymphangiography showed a direct connection between the thoracic duct and the pericardial space (Mask et al. 1990) or a pooling of contrast in the pericardial sac (Goldstein et al. 1969; Yoshida et al. 1969; Chavez et al. 1973; Bhatti et al. 1985). Because it has the advantage of ruling out any obstruction or anomalies of the thoracic duct, it is very useful before considering operative treatment (Yoshida et al. 1969; Modia et al. 1970; Chavez et al. 1973).

The principles of management for chylopericardium are prevention of cardiac tamponade, avoidance of metabolic, nutritional, and immunologic compromise that occurs with a significant chyle leak, and to reduce the likelihood of recurrence. The methods of the management of chylous effusion are as follows: (1) pericardiocentesis for both diagnostic and therapeutic purposes, (2) pericardial window, (3) pericardiectomy, (4) thoracostomy drainage, (5) ligation or resection of the thoracic duct in the lower part of the chest, (6) dietary support with medium or short chain triglyceride and low fat meals (Chavez and Hardy, 1988), (7) pericardioperitoneal (Denver) shunt recently applied in children (Chan et al. 1990). Most of the cases of the chylopericardium have successfully operated regardless of the symptom. Generally the initial treatment of pericardial effusion should consist of open external drainage of the pericardial cavity and a biopsy of the sac (Denfield et al. 1989). The external drainage of the pericardium is a safe procedure and can be performed with a minimum of trauma. In fact, the first reported patient with primary chylopericardium was operated on through a left thoracostomy with the creation of a pleuropericardial window (Grove and Effler, 1954). The pericardial drainage with pericardiocentesis or tube thoracostomy with or without dietary management was effective, but in many cases reaccumulation of the fluid occurred and even in a small number of cases death was also documented (Stratton and Grant, 1958; Miller et al. 1958; Dunn, 1975). The secondary procedure
such as a thoracotomy for ligation of the thoracic duct with or without the creation of the pleuropericardial window or pericardiectomy, can be performed at a later time if the chylopericardium recurs. But considering the results of previous reports, thoracic duct ligation would be the choice of treatment after the diagnosis was confirmed as chylopericardium. Thoracic duct ligation was initially introduced for the management of the chylothorax by Lampson and Conn (1948), who reported a successful case. Thereafter, it was accepted as the standard treatment of the chylothorax of any etiology (Murphy and Piper, 1977). In idiopathic chylopericardium, about two thirds of the cases of primary idiopathic chylopericardium were eventually treated with thoracic duct ligation as well, in which no reaccumulation of chyle was reported (Dunn, 1975; Harada et al. 1982).

Other methods for treatment are pericardiectomy or pericardial window formation without thoracic duct ligation, in which reaccumulation was reported in more than one third of the cases (Sanady and Kovacs, 1973; Dunn, 1975; Ross et al. 1979; Harada et al. 1982; Bendayan et al. 1991). In children three cases of pericardio-peritoneal shunt was reported to be successful (Chan et al. 1990). We can only reemphasize the principles already mentioned before in regard to the treatment of primary idiopathic chylopericardium. To be successful, the thoracic duct should be ligated in a lower part of the thorax and a pericardial window or pericardiectomy should be done concomitantly to ensure adequate drainage.

Although our patient underwent pericardiotomies and the creation of a pleuropericardial window, chylopericardium recurred. Initially and during the follow up period, he was nearly asymptomatic and enjoyed normal everyday activities. The only cause for admission and surgical treatment was a recurrent accumulation of pericardial fluid despite the absence of the symptom. Three months after the last pericardiectomy, he is still asymptomatic and the chest X-ray shows just minimal enlargement of the cardiac shadow. Our plan is to follow him up in the outpatient clinic without further surgical treatment since, in a few asymptomatic chylopericardium of diverse etiology, spontaneous recovery was achieved without surgical treatment in a follow up period of up to four years using only repeated peri-

### SUMMARY

An occurrence of primary idiopathic chylopericardium in a 22-year-old man is described. The asymptomatic nature of this disease in some patients is emphasized to reveal the chronic course of the process. No cause was found for the development of the chylopericardium. However, direct lymphangiography of the lower limbs demonstrated the direct communication between the branches of the thoracic duct and the pericardial sac associated with the development of the chylopericardium. External drainage of the pericardial sac using the subxiphoid route and a pericardio-pleural window formation resulted in temporary success, but chylopericardium recurred 3 months later. Although the ultimate treatment would be a thoracic duct ligation if symptoms appear, no urgent treatment is necessary because he is asymptomatic and the previous operative indication was just a large pericardial effusion without hemodynamic compromise.

### REFERENCES

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